IN THE UNITED STATES DISTRICT COURT
FOR THE DISTRICT OF DELAWARE

NIPPON SHINYAKU CO., LTD., Plaintiff, v. SAREPTA THERAPEUTICS, INC., Defendant.	C.A. No. 21-1015 (GBW) **DEMAND FOR JURY TRIAL**
SAREPTA THERAPEUTICS, INC. and THE UNIVERSITY OF WESTERN AUSTRALIA, Defendant/Counter-Plaintiffs, v. NIPPON SHINYAKU CO., LTD. and NS PHARMA, INC., Plaintiff/Counter-Defendants.	

COUNTER-DEFENDANTS' ANSWER TO
COUNTER-PLAINTIFFS' COUNTERCLAIMS

Counter-Defendants Nippon Shinyaku Co., Ltd. ("Nippon Shinyaku") and NS Pharma, Inc. ("NS Pharma") (collectively, "Counter-Defendants"), by their attorneys, answer the Counterclaims of Counter-Plaintiff Sarepta Therapeutics, Inc. ("Sarepta") and the University of Western Australia ("UWA") (collectively, "Counter-Plaintiffs") and state their affirmative defenses to the Counterclaims asserted against Counter-Defendants. Unless specifically admitted herein each and every allegation in the Counterclaim is denied.

ANSWER TO COUNTERCLAIMS

Responses to Allegations Regarding Nature of the Action

1. Sarepta and UWA assert counterclaims for infringement of U.S. Patent Nos. 9,994,851 ("the '851 patent") (Exhibit A); 10,227,590 ("the '590 patent") (Exhibit B); and 10,266,827 ("the '827 patent") (Exhibit C) (collectively, "the UWA Patents") arising under the patent laws of the United States, 35 U.S.C. § 1 *et seq.* These patent infringement claims arise out of Defendants' unauthorized manufacture, use, sale, offer for sale, and/or importation in the United States of Viltepso, also known as viltolarsen, and Defendants' intentional encouragement of physicians and patients to administer Viltepso.

ANSWER: Counter-Defendants admit that Counter-Plaintiffs' counterclaims purport to assert claims for infringement of the '851 Patent, the '590 Patent, and the '827 Patent. Counter-Defendants deny the remaining allegations in paragraph 1.

2. Sarepta further asserts a counterclaim for declaratory judgment of invalidity of U.S. Patent Nos. 9,708,361 ("the '361 patent"); 10,385,092 ("the '092 patent"); 10,407,461 ("the '461 patent"); 10,487,106 ("the '106 patent"); 10,647,741 ("the '741 patent"); 10,662,217 ("the '217 patent"); and 10,683,322 ("the '322 patent") (collectively, "the NS Patents") arising under the patent laws of the United States, 35 U.S.C. § 1 *et seq.* and under the Declaratory Judgment Act, 28 U.S.C. § 2201 *et seq.*

ANSWER: Counter-Defendants admit that Sarepta's counterclaims purport to assert claims for declaratory judgment of invalidity of the '361 Patent, the '092 Patent, the '461 Patent, the '106 Patent, the '741 Patent, the '217 Patent, and the '322 Patent. Counter-Defendants deny the remaining allegations in paragraph 2.

3. Sarepta further asserts a counterclaim for breach of contract arising under Delaware state law.

ANSWER: Counter-Defendants admit that Sarepta's counterclaims purport to assert a claim for breach of contract arising under Delaware state law. Counter-Defendants deny the remaining allegations in paragraph 3.

Responses to Allegations Regarding the Parties

4. Sarepta is a corporation organized and existing under the laws of the State of Delaware with its principal place of business located at 215 First Street, Cambridge, Massachusetts 02142.

ANSWER: On information and belief, admitted.

5. UWA is a public research university organized and existing under the laws of Australia with its main campus and offices located at 35 Stirling Highway, Crawley, Perth, Western Australia 6009. UWA is the assignee and licensor of the UWA Patents.

ANSWER: On information and belief, admitted.

6. Nippon Shinyaku represents in its Second Amended Complaint that it is a Japanese company with a principal pace of business at 14, Nishinosho-Monguchi-cho, Kisshoin, Minami-ku, Kyoto 601-8550, Japan.

ANSWER: Counter-Defendants admit the allegations in paragraph 6.

7. Nippon Shinyaku represents in its Second Amended Complaint that by virtue of a license agreement with NCNP, Nippon Shinyaku holds the exclusive assertion rights for the NS Patents.

ANSWER: Counter-Defendants admit the allegations in paragraph 7.

8. Upon information and belief, NS Pharma is a corporation organized and existing under the laws of the State of Delaware with its principal place of business at 149 East Ridgewood Ave., Suite 280S, Paramus, NJ 07652. Upon information and belief, NS Pharma is a wholly owned U.S. subsidiary of Nippon Shinyaku. Upon information and belief, NS Pharma is Nippon Shinyaku's U.S. Agent authorized by the FDA to market Viltepso.

ANSWER: Counter-Defendants admit that NS Pharma is a corporation organized and existing under the laws of the State of Delaware. Counter-Defendants further admit that NS Pharma is a wholly-owned subsidiary of Nippon Shinyaku and that NS Pharma is Nippon Shinyaku's U.S. Agent authorized by FDA to market Viltepso. Counter-Defendants admit that NS Pharma has a principal place of business at 140 East Ridgewood Ave., Suite 280S, Paramus, NJ 07652. Counter-Defendants deny the remaining allegations in paragraph 8.

Responses to Allegations Regarding Jurisdiction and Venue

9. There is an actual justiciable controversy between Defendants and Sarepta and UWA concerning Defendants' liability for infringement of the UWA Patents.

ANSWER: Counter-Defendants admit that an actual justiciable controversy exists between Counter-Defendants and Sarepta and UWA regarding Sarepta and UWA's allegations that Counter-Defendants infringe the UWA Patents. Counter-Defendants deny liability for infringement of the UWA Patents and deny the remaining allegations in paragraph 9.

10. Sarepta/UWA's counterclaims against Defendants for infringement of the UWA Patents arise under the patent laws of the United States, 35 U.S.C. § 1 *et seq.*

ANSWER: Paragraph 10 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that Sarepta/UWA's counterclaims against Counter-Defendants for infringement of the UWA Patents arise under the patent laws of the United States, 35 U.S.C. § 1 *et seq.* Counter-Defendants deny the remaining allegations in paragraph 10.

11. This Court has subject matter jurisdiction over the patent infringement counterclaims under 28 U.S.C. §§ 1331 and 1338(a).

ANSWER: Paragraph 11 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that the Court has subject matter jurisdiction over the patent infringement counterclaims under 28 U.S.C. §§ 1331 and 1338(a). Counter-Defendants deny any allegations of infringement of the UWA Patents and deny the remaining allegations in paragraph 11.

12. There is an actual justiciable controversy between Defendants and Sarepta concerning the invalidity of the NS Patents as evidenced by Nippon Shinyaku's allegations in the Second Amended Complaint concerning Sarepta's alleged liability for infringement of the NS Patents.

ANSWER: Paragraph 12 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that there is an actual justiciable

controversy between Counter-Defendants and Sarepta concerning the invalidity of the NS Patents as evidenced by Nippon Shinyaku's allegations in the Second Amended Complaint concerning Sarepta's liability for infringement of the NS Patents. Counter-Defendants deny the remaining allegations in paragraph 12.

13. Sarepta's counterclaim for declaratory judgment of invalidity of the NS Patents arises under the patent laws of the United States, 35 U.S.C. § 1 *et seq.* and under the Declaratory Judgment Act, 28 U.S.C. § 2201 *et seq.*

ANSWER: Paragraph 13 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that Sarepta's counterclaim for declaratory judgment of invalidity of the NS Patents arises under the patent laws of the United States, 35 U.S.C. § 1 *et seq.* and under the Declaratory Judgment Act, 28 U.S.C. § 2201 *et seq.* Counter-Defendants deny the remaining allegations in paragraph 13.

14. This Court has subject matter jurisdiction over the declaratory judgment counterclaim of invalidity of the NS Patents under 28 U.S.C. §§ 1331, 1338(a), 2201, and 2202.

ANSWER: Paragraph 14 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that this Court has subject matter jurisdiction over the declaratory judgment counterclaim of invalidity of the NS Patents under 28 U.S.C. §§ 1331, 1338(a), 2201, and 2202. Counter-Defendants deny the remaining allegations in paragraph 14.

15. There is an actual justiciable controversy between Nippon Shinyaku and Sarepta concerning Nippon Shinyaku's breach of contract.

ANSWER: Paragraph 15 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that Sarepta's counterclaims purport to assert a breach of contract claim. Counter-Defendants deny the remaining allegations in paragraph 15.

16. Sarepta's breach of contract counterclaim arises under Delaware state law.

ANSWER: Paragraph 16 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that Sarepta's counterclaims purport to assert a breach of contract claim under Delaware law. Counter-Defendants deny the remaining allegations in paragraph 16.

17. This Court has subject matter jurisdiction over the breach of contract counterclaim under 28 U.S.C. §§ 1332(a) and 1367(a).

ANSWER: Paragraph 17 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that Sarepta's counterclaims purport to assert a breach of contract claim. Counter-Defendants deny the remaining allegations in paragraph 17.

18. Personal jurisdiction is proper over Nippon Shinyaku at least because Nippon Shinyaku has commenced this action and thus submitted to this Court's personal jurisdiction.

ANSWER: Paragraph 18 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that personal jurisdiction is proper over Nippon Shinyaku for purposes of this action only at least because Nippon Shinyaku has commenced this action and thus submitted to this Court's personal jurisdiction. Counter-Defendants deny the remaining allegations in paragraph 18.

19. Upon information and belief, personal jurisdiction is proper over NS Pharma, a Delaware corporation, at least because it has committed acts of infringement of the UWA Patents in Delaware by offering to sell and selling Viltepso (viltolarsen) in the State of Delaware. In addition, upon information and belief, Nippon Shinyaku conferred with, and coordinated with, NS Pharma in bringing this action and thus NS Pharma has consented to this Court's personal jurisdiction.

ANSWER: Paragraph 19 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that personal jurisdiction is proper over NS Pharma for purposes of this action only. Counter-Defendants deny the remaining

allegations in paragraph 19.

20. Upon information and belief, Nippon Shinyaku directly or through its agents including its wholly owned U.S. subsidiary NS Pharma, manufactures, markets, offers to sell, sells, and/or distributes Viltepso (viltolarsen) in the State of Delaware and elsewhere in the United States, and Viltepso is prescribed by physicians practicing in Delaware and elsewhere in the United States, is available at pharmacies or medical facilities located within Delaware and elsewhere in the United States, and/or is used by patients in, and/or residents of, Delaware and elsewhere in the United States.

ANSWER: Counter-Defendants admit that Nippon Shinyaku directly or through its agents and other third parties manufactures, markets, offers to sell, sells, and/or distributes Viltepso (viltolarsen) in the United States. Counter-Defendants also admit that Viltepso is prescribed by physicians practicing in the United States, is available at pharmacies or medical facilities in the United States, and/or is used by patients in, and/or residents of, Delaware and elsewhere in the United States. Counter-Defendants deny the remaining allegations in paragraph 20.

21. Venue is proper in this Court pursuant to 28 U.S.C. §§ 1391(c)(3) and 1400(b).

ANSWER: Paragraph 21 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that venue is proper in this Court for purposes of this action only. Counter-Defendants deny the remaining allegations in paragraph 21.

Responses to Allegations Regarding the UWA Patents

22. On June 12, 2018, the USPTO issued the '851 patent, entitled "Antisense Oligonucleotides for Inducing Exon Skipping and Methods of Use Thereof." The '851 patent is assigned to The University of Western Australia. A copy of the '851 patent is attached hereto as Exhibit A. The '851 patent is fully maintained and is valid and enforceable. Sarepta has exclusive rights to the '851 patent for the treatment of muscular dystrophies and the right to enforce the '851 patent.

ANSWER: Counter-Defendants admit that the '851 patent is entitled "Antisense Oligonucleotides for Inducing Exon Skipping and Methods of Use Thereof" and states that it issued on June 12, 2018. Counter-Defendants further admit that the face of the '851 patent lists the Assignee as the University of Western Australia and that Sarepta's Exhibit A purports to be a

copy of the ’851 patent. Counter-Defendants deny that the ’851 patent is valid and enforceable. Counter-Defendants are without knowledge or information sufficient to form a belief as to the truth of the remaining allegations in paragraph 22 and therefore deny the same.

23. On March 12, 2019, the USPTO issued the ‘590 patent, entitled “Antisense Oligonucleotides for Inducing Exon Skipping and Methods of Use Thereof.” The ‘590 patent is assigned to The University of Western Australia. A copy of the ‘590 patent is attached hereto as Exhibit B. The ‘590 patent is fully maintained and is valid and enforceable. Sarepta has exclusive rights to the ‘590 patent for the treatment of muscular dystrophies and the right to enforce the ‘590 patent.

ANSWER: Counter-Defendants admit that the ’590 patent is entitled “Antisense Oligonucleotides for Inducing Exon Skipping and Methods of Use Thereof” and states that it issued on March 12, 2019. Counter-Defendants further admit that the face of the ’590 patent lists the Assignee as the University of Western Australia and that Sarepta’s Exhibit B purports to be a copy of the ’590 patent. Counter-Defendants deny that the ’590 patent is valid and enforceable. Counter-Defendants are without knowledge or information sufficient to form a belief as to the truth of the remaining allegations in paragraph 23 and therefore deny the same.

24. On April 23, 2019, the USPTO issued the ‘827 patent, entitled “Antisense Oligonucleotides for Inducing Exon Skipping and Methods of Use Thereof.” The ‘827 patent is assigned to The University of Western Australia. A copy of the ‘827 patent is attached hereto as Exhibit C. The ‘827 patent is fully maintained and is valid and enforceable. Sarepta has exclusive rights to the ‘827 patent for the treatment of muscular dystrophies and the right to enforce the ‘827 patent.

ANSWER: Counter-Defendants admit that the ’827 patent is entitled “Antisense Oligonucleotides for Inducing Exon Skipping and Methods of Use Thereof” and states that it issued on April 23, 2019. Counter-Defendants further admit that the face of the ’827 patent lists the Assignee as the University of Western Australia and that Sarepta’s Exhibit C purports to be a copy of the ’827 patent. Counter-Defendants deny that the ’827 patent is valid and enforceable. Counter-Defendants are without knowledge or information sufficient to form a belief as to the truth of the remaining allegations in paragraph 24 and therefore deny the same.

25. The UWA Patents are listed in the U.S. Food and Drug Administration's ("FDA") *Approved Drug Products with Therapeutic Equivalence Evaluations* ("the Orange Book") for New Drug Application ("NDA") No. 211970 for Sarepta's Vyondys 53® product, also known as golodirsen. Each of the UWA Patents covers, *inter alia*, an antisense oligonucleotide of 20 to 31 bases wherein a base sequence comprises at least 12 consecutive bases of SEQ ID NO: 195 disclosed in the UWA Patents, in which uracil bases are thymine bases, and a method of using it for the treatment of Duchenne Muscular Dystrophy ("DMD") in patients who have a mutation of the DMD gene that is amenable to exon 53 skipping.

ANSWER: Counter-Defendants admit that the UWA Patents are listed in the U.S. Food and Drug Administration's ("FDA") Approved Drug Products with Therapeutic Equivalence Evaluations ("the Orange Book") for New Drug Application ("NDA") No. 211970 for Sarepta's Vyondys 53® product, also known as golodirsen. Counter-Defendants further admit that each of the claims in the UWA Patents claims, inter alia, an antisense oligonucleotide of 20 to 31 bases, wherein the base sequence comprises at least 12 consecutive bases of SEQ ID NO: 195 in which uracil bases are thymine bases. Counter-Defendants further admit that the claims of the '827 patent claim a method for treating a patient with DMD in need thereof who has a mutation of the DMD gene that is amenable to exon 53 skipping. Counter-Defendants deny the remaining allegations in paragraph 25.

Responses to Allegations Regarding Defendants' Infringing Product[1]

26. Upon information and belief, Defendants' product, Viltepso (viltolarsen), is a morpholino antisense oligonucleotide comprising a base sequence that is 100% complementary to consecutive bases of a target region of exon 53 of the human dystrophin pre-mRNA. Viltepso (viltolarsen) Highlights of Prescribing Information (Aug. 2020),[2] § 11; *see also* Viltepso (viltolarsen) Highlights of Prescribing Information (Mar. 2021)[3]. Viltepso contains 21 bases and

[1] Counter-Defendants have adopted the headings as provided in Counter-Plaintiff's Counterclaims for ease of reference only. Counter-Defendants do not admit any allegation found in any of the headings and deny that their product is "infringing."

[2] Viltepso (viltolarsen) Highlights of Prescribing Information (Rev. Aug. 2020), https://www.accessdata.fda.gov/drugsatfda_docs/label/2020/212154Orig1s000lbl.pdf (last visited Jan. 28, 2022).

[3] Viltepso (viltolarsen) Highlights of Prescribing Information (Rev. Mar. 2021), https://www.viltepso.com/prescribing-information (last visited Jan. 28, 2022).

CCTCCGGTTCTGAAGGTGTTC as the base sequence. Viltepso (viltolarsen) Highlights of Prescribing Information (Mar. 2021), § 11.

ANSWER: Counter-Defendants admit that § 11 of the Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of August 2020 and with a revision date of March 2021 states "Viltolarsen is an antisense oligonucleotide of the phosphorodiamidate morpholino oligomer (PMO) subclass." Counter-Defendants also admit that § 11 of the Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of August 2020 and with a revision date of March 2021 states "Viltolarsen contains 21 linked subunits." Counter-Defendants further admit that the sequence of bases of Viltepso from the 5' end to the 3' end is CCTCCGGTTCTGAAGGTGTTC. Counter-Defendants deny the remaining allegations in paragraph 26.

27. Upon information and belief, Viltepso induces exon 53 skipping in a human dystrophin pre-mRNA. *Id.* § 12.1.

ANSWER: Counter-Defendants admit that § 12.1 of the Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of August 2020 and with a revision date of March 2021 states "VILTEPSO is designed to bind to exon 53 of dystrophin pre-mRNA resulting in exclusion of this exon during mRNA processing in patients with genetic mutations that are amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 27.

28. Upon information and belief, Viltepso is administered to DMD patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping and induces skipping of exon 53 of dystrophin pre-mRNA. *Id.* §§ 1, 12.1. Defendants' label for Viltepso has encouraged and continues to encourage such use.

ANSWER: Counter-Defendants admit that Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of August 2020 and with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy

(DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping," and § 12.1 states "VILTEPSO is designed to bind to exon 53 of dystrophin pre-mRNA resulting in exclusion of this exon during mRNA processing in patients with genetic mutations that are amenable to exon 53 skipping. Exon 53 skipping is intended to allow for production of an internally truncated dystrophin protein in patients with genetic mutations that are amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 28.

29. Upon information and belief, Defendants conducted pre-clinical and clinical development of Viltepso (viltolarsen), including clinical trials, to generate data in support of their submission of an NDA with the FDA for Viltepso (viltolarsen).

ANSWER: Counter-Defendants admit that Nippon Shinyaku conducted pre-clinical and clinical development of Viltepso (viltolarsen), including clinical trials, to generate data in support of the submission of an NDA with the FDA for Viltepso (viltolarsen). Counter-Defendants deny that NS Pharma was involved in the pre-clinical development of Viltepso. Counter-Defendants deny the remaining allegations in paragraph 29.

30. Upon information and belief, on October 2, 2019, Nippon Shinyaku announced that it had submitted a rolling NDA for Viltepso (viltolarsen) with the FDA. Nippon Shinyaku News Release (Oct. 2, 2019).[4]

ANSWER: Counter-Defendants admit that the article cited in Counter-Plaintiffs' counterclaims titled "U.S. FDA Submission of New Drug Application for NS-065/NCNP-01 (viltolarsen)" dated October 2, 2019 states that Nippon Shinyaku "announced that it has completed the submission of its rolling New Drug Application (NDA) to the U.S. Food and Drug Administration (FDA) for NS-065/NCNP-01 (viltolarsen)." Counter-Defendants deny the remaining allegations in paragraph 30.

[4] Nippon Shinyaku Press Release (Oct. 2, 2019), https://www.nippon-shinyaku.co.jp/file/download.php?file_id=3838 (last visited Jan. 28, 2022).

31. On August 12, 2020, the FDA announced it had granted accelerated approval to Viltepso for the treatment of DMD in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping. FDA News Release (Aug. 12, 2020).[5]

ANSWER: Counter-Defendants admit that the article cited in Counter-Plaintiffs' counterclaims titled "FDA Approves Targeted Treatment for Rare Duchenne Muscular Dystrophy Mutation" dated August 12, 2020 states that "[t]oday, the U.S. Food and Drug Administration granted accelerated approval to Viltepso (viltolarsen) injection for the treatment of Duchenne muscular dystrophy (DMD) in patients wo have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 31.

32. Upon information and belief, Nippon Shinyaku announced that NS Pharma, a wholly owned U.S. subsidiary of Nippon Shinyaku, had launched Viltepso for commercial sales in the United States as of August 19, 2020. Nippon Shinyaku News Release (Aug. 20, 2020).[6] Upon information and belief, NS Pharma is Nippon Shinyaku's U.S. Agent authorized by the FDA to market Viltepso. *Id.*

ANSWER: Counter-Defendants admit that the article cited in Counter-Plaintiffs' counterclaims titled "VILTEPSO™ (viltolarsen) injection Now Commercially Available in the U.S." dated August 20, 2020 states that "Nippon Shinyaku Co., Ltd. . . . announced today that NS Pharma, Inc. . . . a wholly owned subsidiary of Nippon Shinyaku made VILTEPSO™ (viltolarsen) now available for commercial sales in the United States market as of August 19 (EST)." Counter-Defendants further admit that NS Pharma is Nippon Shinyaku's U.S. Agent authorized by the FDA to market Viltepso. Counter-Defendants deny the remaining allegations in paragraph 32.

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[5] FDA News Release (Aug. 12, 2020), https://www.fda.gov/news-events/press-announcements/fda-approves-targeted-treatment-rare-duchenne-muscular-dystrophy-mutation (last visited Jan. 28, 2022).

[6] Nippon Shinyaku Press Release (Aug. 20, 2020), https://www.nippon-shinyaku.co.jp/file/download.php?file_id=3868 (last visited Jan. 28, 2022).

33. Upon information and belief, since at least August 2020, Defendants have encouraged physicians to treat DMD patients by administering Viltepso to induce skipping of exon 53 of dystrophin pre-mRNA including through their labels for Viltepso. Defendants have also facilitated pricing and reimbursement of Viltepso in the United States.

ANSWER: Paragraph 33 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping. Counter-Defendants deny the remaining allegations in paragraph 33.

Responses to Allegations Regarding Defendants' Awareness of the UWA Patents

34. Upon information and belief, Defendants have been familiar with and knew of the UWA Patents prior to this litigation. Upon information and belief, Defendants believed prior to this litigation that one or more claims of the UWA Patents covered Viltepso (viltolarsen). When the UWA Patents issued in 2018 and 2019, for example, Defendants' NDA seeking marketing approval for viltolarsen was under regulatory review in the United States. Upon information and belief, Defendants became aware of the UWA Patents after the UWA Patents were submitted for listing in the FDA Orange Book for Vyondys 53® in December 2019. Upon information and belief, Defendants expected that their Viltepso (viltolarsen) product, if approved, would compete directly with Sarepta's Vyondys 53® (golodirsen) product. Upon information and belief, Defendants learned of the UWA Patents through their efforts to research and/or monitor third-party U.S. patents that could potentially interfere with their ability to market Viltepso (viltolarsen) in the United States.

ANSWER: Counter-Defendants admit that they were aware of the UWA Patents by at least September 2019. Counter-Defendants further admit that the UWA Patents include claims aimed at capturing VILTEPSO. Counter-Defendants admit that the NDA seeking marketing approval for viltolarsen was under regulatory review in the United States in 2018 and 2019. Counter-Defendants further admit that Nippon Shinyaku and Sarepta are direct competitors in certain markets for antisense oligonucleotide-based therapies for the treatment of DMD. Counter-Defendants deny the remaining allegations in paragraph 34.

35. Sarepta and Nippon Shinyaku entered into a Mutual Confidentiality Agreement ("MCA") effective June 1, 2020. Upon information and belief, Defendants were already aware of the UWA Patents when Sarepta and Nippon Shinyaku began business discussions under the MCA in June 2020.

ANSWER: Counter-Defendants admit that Sarepta and Nippon Shinyaku entered into a Mutual Confidentiality Agreement ("MCA") effective June 1, 2020. Counter-Defendants further admit that they were aware of the UWA Patents at least as of September 2019. Counter-Defendants deny the remaining allegations in paragraph 35.

RESPONSES TO ALLEGATIONS REGARDING COUNTERCLAIM I
(Infringement of the '851 Patent)

36. Sarepta and UWA reallege each of the foregoing Paragraphs 1-35 as if fully set forth herein.

ANSWER: Counter-Defendants incorporate by reference their answers to the allegations of Paragraphs 1-35 as if fully set forth herein.

37. Sarepta and UWA incorporate by reference Sarepta's answers and responses to Nippon Shinyaku's Second Amended Complaint ("SAC") as if fully set forth herein.

ANSWER: Counter-Defendants incorporate by reference the Second Amended Complaint as if fully set forth herein.

38. Claim 1 of the '851 patent recites:

> An antisense oligonucleotide of 20 to 31 bases comprising a base sequence that is 100% complementary to consecutive bases of a target region of exon 53 of the human dystrophin pre-mRNA, wherein the target region is within annealing site H53A(+23+47) and annealing site H53A(+39+69), wherein the base sequence comprises at least 12 consecutive bases of CUG AAG GUG UUC UUG UAC UUC AUC C (SEQ ID NO: 195), in which uracil bases are thymine bases, wherein the antisense oligonucleotide is a morpholino antisense oligonucleotide, and wherein the antisense oligonucleotide induces exon 53 skipping; or a pharmaceutically acceptable salt thereof.

ANSWER: Counter-Defendants admit that paragraph 38 quotes claim 1 of the '851 patent.

39. Upon information and belief, Viltepso satisfies each element of at least claim 1 of the '851 patent.

ANSWER: Counter-Defendants deny the allegations in paragraph 39.

40. Viltolarsen is an antisense oligonucleotide of the phosphorodiamidate morpholino oligomer (PMO) subclass. *See* Viltepso (viltolarsen) Highlights of Prescribing Information (Rev. Mar. 2021), § 11. Viltolarsen contains 21 linked subunits. *Id.* The base sequence of viltolarsen is CCTCCGGTTCTGAAGGTGTTC, which includes CTGAAGGTGTTC as 12 consecutive bases. *Id.*

ANSWER: Counter-Defendants admit that § 11 of the Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states "Viltolarsen is an antisense oligonucleotide of the phosphorodiamidate morpholino oligomer (PMO) subclass." Counter-Defendants also admit that § 11 of the Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states "Viltolarsen contains 21 linked subunits." Counter-Defendants further admit that the sequence of bases of Viltepso from the 5' end to the 3' end is CCTCCGGTTCTGAAGGTGTTC. Counter-Defendants deny the remaining allegations in paragraph 40.

41. Viltepso is designed to bind to exon 53 of dystrophin pre-mRNA resulting in exclusion of this exon during mRNA processing in patients with genetic mutations that are amenable to exon 53 skipping. *Id.* § 12.1.

ANSWER: Counter-Defendants admit that Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states in § 12.1 that "VILTEPSO is designed to bind to exon 53 of dystrophin pre-mRNA resulting in exclusion of this exon during mRNA processing in patients with genetic mutations that are amenable to exon 53 skipping. Exon 53 skipping is intended to allow for production of an internally truncated dystrophin protein in patients with genetic mutations that are amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 41.

42. Viltepso is indicated for the treatment of DMD in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping. *Id.* § 1.

ANSWER: Counter-Defendants admit that Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 42.

43. Upon information and belief, Defendants have directly infringed and continue to directly infringe at least claim 1 of the '851 patent, either literally or under the doctrine of equivalents, by engaging in the commercial manufacture, use, offer for sale, sale, and/or importation of Viltepso in the United States in violation of 35 U.S.C. § 271(a).

ANSWER: Counter-Defendants deny the allegations in paragraph 43.

44. Upon information and belief, Defendants knew or should have known of the existence of the '851 patent at least as of the date of its issuance. For example, upon information and belief, Defendants learned of the '851 patent through their monitoring of third-party patent rights, including the UWA Patents, and their efforts to obtain approval for, and market, viltolarsen in the United States. Upon information and belief, Defendants were aware of the '851 patent before June 2020 when Nippon Shinyaku and Sarepta began business discussions under the MCA.

ANSWER: Counter-Defendants admit that they were aware of the '851 patent by at least September 2019. Counter-Defendants deny the remaining allegations in paragraph 44.

45. Viltepso is approved for use in the treatment of DMD in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping. *See* Viltepso (viltolarsen) Highlights of Prescribing Information (Rev. Mar. 2021), § 1. Upon information and belief, Viltepso has no substantial non-infringing uses, and Defendants know that Viltepso is especially made or especially adapted for use in infringement of the '851 patent.

ANSWER: Counter-Defendants admit that Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 45.

46. Upon information and belief, Defendants' sale, offer for sale, and/or distribution of Viltepso includes labeling indicating that it is approved by the FDA to treat DMD in patients who have a confirmed mutation of the dystrophin gene that is amenable to exon 53 skipping and instructs physicians and patients to use Viltepso to treat DMD. *Id.* Upon information and belief, Defendants specifically intend that Viltepso be used to treat DMD with the knowledge that it would infringe the '851 patent.

ANSWER: Counter-Defendants admit that Viltepso is sold, offered for sale, and/or distributed in the United States and that its Highlights of Prescribing Information with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 46.

47. Upon information and belief, Defendants have induced and/or contributed to and continue to induce and/or contribute to infringement of at least claim 1 of the '851 patent, either literally or under the doctrine of equivalents, in violation of 35 U.S.C. §§ 271(b) and/or (c).

ANSWER: Counter-Defendants deny the allegations in paragraph 47.

48. Upon information and belief, Defendants' infringement of the '851 patent has been willful and continues to be willful, entitling Sarepta and UWA to enhanced damages pursuant to 35 U.S.C. § 284. Upon information and belief, Defendants knew or should have known that their conduct related to the development, manufacture, use, offer for sale, sale, and/or importation of Viltepso constituted an unreasonable risk of infringement of at least claim 1 of the '851 patent.

ANSWER: Counter-Defendants deny the allegations in paragraph 48.

49. This case is exceptional and Sarepta and UWA are entitled to attorneys' fees and costs incurred in prosecuting this action pursuant to 35 U.S.C. § 285.

ANSWER: Counter-Defendants deny the allegations in paragraph 49.

RESPONSES TO ALLEGATIONS REGARDING COUNTERCLAIM II
(Infringement of the '590 Patent)

50. Sarepta and UWA reallege each of the foregoing Paragraphs 1-49 as if fully set forth herein.

ANSWER: Counter-Defendants incorporate by reference their answers to the allegations of Paragraphs 1-49 as if fully set forth herein.

51. Sarepta and UWA incorporate by reference Sarepta's answers and responses to Nippon Shinyaku's Second Amended Complaint as if fully set forth herein.

ANSWER: Counter-Defendants incorporate by reference the Second Amended Complaint as if fully set forth herein.

52. Claim 1 of the '590 patent recites:

> An antisense oligonucleotide of 20 to 31 bases comprising a base sequence that is 100% complementary to consecutive bases of a target region of exon 53 of the human dystrophin pre-mRNA, wherein the base sequence comprises at least 12 consecutive bases of CUG AAG GUG UUC UUG UAC UUC AUC C (SEQ ID NO: 195), in which uracil bases are thymine bases, wherein the antisense oligonucleotide is a morpholino antisense oligonucleotide, and wherein the antisense oligonucleotide induces exon 53 skipping; or a pharmaceutically acceptable salt thereof.

ANSWER: Counter-Defendants admit that paragraph 52 quotes claim 1 of the '590 patent.

53. Upon information and belief, Viltepso satisfies each element of at least claim 1 of the '590 patent.

ANSWER: Counter-Defendants deny the allegations in paragraph 53.

54. Viltolarsen is an antisense oligonucleotide of the phosphorodiamidate morpholino oligomer (PMO) subclass. *See* Viltepso (viltolarsen) Highlights of Prescribing Information (Rev. Mar. 2021), § 11. Viltolarsen contains 21 linked subunits. *Id.* The base sequence of viltolarsen is CCTCCGGTTCTGAAGGTGTTC, which includes CTGAAGGTGTTC as 12 consecutive bases. *Id.*

ANSWER: Counter-Defendants admit that § 11 of the Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states "Viltolarsen is an antisense oligonucleotide of the phosphorodiamidate morpholino oligomer (PMO) subclass." Counter-

Defendants also admit that § 11 of the Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states "Viltolarsen contains 21 linked subunits." Counter-Defendants further admit that the sequence of bases of Viltepso from the 5' end to the 3' end is CCTCCGGTTCTGAAGGTGTTC. Counter-Defendants deny the remaining allegations in paragraph 54.

55. Viltepso is designed to bind to exon 53 of dystrophin pre-mRNA resulting in exclusion of this exon during mRNA processing in patients with a genetic mutation that is amenable to exon 53 skipping. *Id.* § 12.1.

ANSWER: Counter-Defendants admit that Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states in § 12.1 that "VILTEPSO is designed to bind to exon 53 of dystrophin pre-mRNA resulting in exclusion of this exon during mRNA processing in patients with genetic mutations that are amenable to exon 53 skipping. Exon 53 skipping is intended to allow for production of an internally truncated dystrophin protein in patients with genetic mutations that are amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 55.

56. Viltepso is indicated for the treatment of DMD in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping. *Id.* § 1.

ANSWER: Counter-Defendants admit that Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 56.

57. Upon information and belief, Defendants have directly infringed and continue to directly infringe at least claim 1 of the '590 patent, either literally or under the doctrine of equivalents, by engaging in the commercial manufacture, use, offer for sale, sale, and/or importation of Viltepso in the United States in violation of 35 U.S.C. § 271(a).

ANSWER: Counter-Defendants deny the allegations in paragraph 57.

58. Upon information and belief, Defendants knew or should have known of the existence of the '590 patent at least as of the date of its issuance. For example, upon information and belief, Defendants learned of the '590 patent through their monitoring of third-party patent rights, including the UWA Patents, and their efforts to obtain approval for, and market, viltolarsen in the United States. Upon information and belief, Defendants were aware of the '590 patent before June 2020 when Nippon Shinyaku and Sarepta began business discussions under the MCA.

ANSWER: Counter-Defendants admit that they were aware of the '590 patent by at least September 2019. Counter-Defendants deny the remaining allegations in paragraph 58.

59. Viltepso is approved for use in the treatment of DMD in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping. *See* Viltepso (viltolarsen) Highlights of Prescribing Information (Rev. Mar. 2021), § 1. Upon information and belief, viltolarsen has no substantial non-infringing uses, and Defendants know that viltolarsen is especially made or especially adapted for use in infringement of the '590 patent.

ANSWER: Counter-Defendants admit that Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 59.

60. Upon information and belief, Defendants' sale, offer for sale, and/or distribution of Viltepso includes labeling indicating that it is approved by the FDA to treat DMD in patients who have a confirmed mutation of the dystrophin gene that is amenable to exon 53 skipping and instructs physicians and patients to use Viltepso to treat DMD. *Id.* Upon information and belief, Defendants specifically intend that Viltepso be used to treat DMD with the knowledge that it would infringe the '590 patent.

ANSWER: Counter-Defendants admit that Viltepso is sold, offered for sale, and/or distributed in the United States and that its Highlights of Prescribing Information with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 60.

61. Upon information and belief, Defendants have induced and/or contributed to and continue to induce and/or contribute to infringement of at least claim 1 of the '590 patent, either literally or under the doctrine of equivalents, in violation of 35 U.S.C. §§ 271(b) and/or (c).

ANSWER: Counter-Defendants deny the allegations in paragraph 61.

62. Upon information and belief, Defendants' infringement of the '590 patent has been willful and continues to be willful, entitling Sarepta and UWA to enhanced damages pursuant to 35 U.S.C. § 284. Upon information and belief, Defendants knew or should have known that their conduct related to the development, manufacture, use, offer for sale, sale, and/or importation of Viltepso constituted an unreasonable risk of infringement of at least claim 1 of the '590 patent.

ANSWER: Counter-Defendants deny the allegations in paragraph 62.

63. This case is exceptional and Sarepta and UWA are entitled to attorneys' fees and costs incurred in prosecuting this action pursuant to 35 U.S.C. § 285.

ANSWER: Counter-Defendants deny the allegations in paragraph 63.

RESPONSES TO ALLEGATIONS REGARDING COUNTERCLAIM III
(Infringement of the '827 Patent)

64. Sarepta and UWA reallege each of the foregoing Paragraphs 1-63 as if fully set forth herein.

ANSWER: Counter-Defendants incorporate by reference their answers to the allegations of Paragraphs 1-63 as if fully set forth herein.

65. Sarepta and UWA incorporate by reference Sarepta's answer and responses to Nippon Shinyaku's Second Amended Complaint as if fully set forth herein.

ANSWER: Counter-Defendants incorporate by reference the Second Amended Complaint as if fully set forth herein.

66. Claim 1 of the '827 patent recites:

> A method for treating a patient with Duchenne muscular dystrophy (DMD) in need thereof who has a mutation of the DMD gene that is amenable to exon 53 skipping, comprising administering to the patient an antisense oligonucleotide of 20 to 31 bases comprising a base sequence that is 100% complementary to consecutive bases of a target region of exon 53 of the human dystrophin pre-mRNA, wherein the base sequence comprises at least 12 consecutive bases of CUG AAG GUG UUC UUG UAC UUC AUC C (SEQ ID NO: 195), in which uracil bases are thymine bases, wherein the antisense oligonucleotide is a morpholino antisense

> oligonucleotide, and wherein the antisense oligonucleotide induces exon 53 skipping; or a pharmaceutically acceptable salt thereof.

ANSWER: Counter-Defendants admit that paragraph 66 quotes claim 1 of the '827 patent.

67. Upon information and belief, the use of Viltepso satisfies each element of, and directly infringes, either literally or under the doctrine of equivalents, at least claim 1 of the '827 patent.

ANSWER: Counter-Defendants deny the allegations in paragraph 67.

68. Viltepso is indicated for the treatment of DMD in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping. *See* Viltepso (viltolarsen) Highlights of Prescribing Information (Rev. Mar. 2021), § 1.

ANSWER: Counter-Defendants admit that Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 68.

69. Upon information and belief, Defendants knew or should have known of the existence of the '827 patent at least as of the date of its issuance. For example, upon information and belief, Defendants learned of the '827 patent through their monitoring of third-party patent rights, including the UWA Patents, and their efforts to obtain approval for, and market, viltolarsen in the United States. Upon information and belief, Defendants were aware of the '827 patent before June 2020 when Nippon Shinyaku and Sarepta began business discussions under the MCA.

ANSWER: Counter-Defendants admit that they were aware of the '827 patent by at least September 2019. Counter-Defendants deny the remaining allegations in paragraph 69.

70. Viltepso is approved for use in the treatment of DMD in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping. *Id.* Upon information and belief, viltolarsen has no substantial non-infringing uses, and Defendants know that viltolarsen is especially made or especially adapted for use in infringement of the '827 patent.

ANSWER: Counter-Defendants admit that Viltepso (viltolarsen) Highlights of Prescribing Information with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a

confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 70.

71. Upon information and belief, Defendants' sale, offer for sale, and/or distribution of Viltepso includes labeling indicating that it is approved by the FDA to treat DMD in patients who have a confirmed mutation of the dystrophin gene that is amenable to exon 53 skipping and instructs physicians and patients to use Viltepso to treat DMD. *Id.* Upon information and belief, Defendants specifically intend that Viltepso be used to treat DMD with the knowledge that it would infringe the '827 patent.

ANSWER: Counter-Defendants admit that Viltepso is sold, offered for sale, and/or distributed in the United States and that its Highlights of Prescribing Information with a revision date of March 2021 states in § 1 that "VILTEPSO is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping." Counter-Defendants deny the remaining allegations in paragraph 71.

72. Upon information and belief, Defendants have induced and/or contributed to and continue to induce and/or contribute to infringement of at least claim 1 of the '827 patent, either literally or under the doctrine of equivalents, in violation of 35 U.S.C. §§ 271(b) and/or (c).

ANSWER: Counter-Defendants deny the allegations in paragraph 72.

73. Upon information and belief, Defendants' infringement of the '827 patent has been willful and continues to be willful, entitling Sarepta and UWA to enhanced damages pursuant to 35 U.S.C. § 284. Upon information and belief, Defendants knew or should have known that their conduct related to the development, manufacture, use, offer for sale, sale, and/or importation of Viltepso constituted an unreasonable risk of infringement of at least claim 1 of the '827 patent.

ANSWER: Counter-Defendants deny the allegations in paragraph 73.

74. This case is exceptional and Sarepta and UWA are entitled to attorneys' fees and costs incurred in prosecuting this action pursuant to 35 U.S.C. § 285.

ANSWER: Counter-Defendants deny the allegations in paragraph 74.

RESPONSES TO ALLEGATIONS REGARDING COUNTERCLAIM IV
(Declaration of Invalidity of the NS Patents)

75. Sarepta realleges each of the foregoing Paragraphs 1-74 as if fully set forth herein.

ANSWER: Counter-Defendants incorporate by reference their answers to the allegations of Paragraphs 1-74 as if fully set forth herein.

76. Sarepta incorporates by reference its answers and responses to Nippon Shinyaku's Second Amended Complaint.

ANSWER: Counter-Defendants incorporate by reference the Second Amended Complaint as if fully set forth herein.

77. Each claim of the NS Patents is invalid for failure to comply with one or more requirements of the patent laws of the United States, including without limitation, 35 U.S.C. §§ 101, 102, 103, 112, and/or obviousness-type double patenting, and the rules, regulations, and laws pertaining thereto.

ANSWER: Counter-Defendants deny the allegations in paragraph 77.

78. By way of example, the claims of the NS Patents are invalid under 35 U.S.C. §§ 102 and/or 103 in view of Popplewell, L.J., *Comparative Analysis of Antisense Oligonucleotide Sequences Targeting Exon 53 of the Human DMD Gene: Implications for Future Clinical Trials*, Neuromuscular Disorders 20:102–110 (2010) ("Popplewell") and Sazani, P., *Safety Pharmacology and Genotoxicity Evaluation of AVI-4658*, Int'l J. Toxicology 29(2):143–156 (2010) ("Sazani"), alone or in combination with other prior art, for at least the reasons set forth in Sarepta's IPR Petitions challenging the NS Patents. In granting Sarepta's IPR Petitions challenging all claims of all seven NS Patents, for example, the Patent Trial and Appeal Board found Sarepta's arguments and evidence of unpatentability persuasive, concluding in each institution decision that Sarepta "has demonstrated a reasonable likelihood of success in proving that the challenged claims of the [patent] are unpatentable." *See Sarepta Therapeutics, Inc. v. Nippon Shinyaku Co., Ltd.*, Decisions Granting Institution in IPR2021-01134, Paper No. 20 (Jan. 12, 2022); IPR2021-01135, Paper No. 20 (Jan. 12, 2022); IPR2021-01136, Paper No. 19 (Jan. 13, 2022); IPR2021-01137, Paper No. 18 (Jan. 13, 2022); IPR2021-01138, Paper No. 18 (Jan. 13, 2022); IPR2021-01139, Paper No. 18 (Jan. 13, 2022); and IPR2021-01140, Paper No. 18 (Jan. 12, 2022).

ANSWER: Counter-Defendant admits that the Patent Trial and Appeal Board stated in the Institution Decisions for the IPR Petitions that Sarepta "has demonstrated a reasonable likelihood of success in proving that the challenged claims of the [patent] are unpatentable." Counter-Defendants deny the remaining allegations in paragraph 78.

79. An actual case or controversy exists between Sarepta and Defendants as to whether the claims of the NS Patents are invalid.

ANSWER: Paragraph 79 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that there is an actual case or controversy between Counter-Defendants and Sarepta concerning the invalidity of the NS Patents. Counter-Defendants deny the remaining allegations in paragraph 79.

80. Sarepta is entitled to a declaratory judgment that the claims of the NS Patents are invalid.

ANSWER: Counter-Defendants deny the allegations of paragraph 80.

81. This case is exceptional and Sarepta is entitled to attorneys' fees and costs incurred in prosecuting this action pursuant to 35 U.S.C. § 285.

ANSWER: Counter-Defendants deny the allegations of paragraph 81.

RESPONSES TO ALLEGATIONS REGARDING COUNTERCLAIM V
(Breach of Contract)

82. Sarepta realleges each of the foregoing Paragraphs 1-81 as if fully set forth herein.

ANSWER: Counter-Defendants incorporate by reference their answers to the allegations of Paragraphs 1-81 as if fully set forth herein.

83. Sarepta incorporates by reference its answers and responses in Sarepta's Answer and Counterclaims to Nippon Shinyaku's Second Amended Complaint as fully set forth herein.

ANSWER: Counter-Defendants incorporate by reference the Second Amended Complaint as if fully set forth herein.

84. Sarepta asserts a claim for breach of contract arising under Delaware state law. This Court has subject matter jurisdiction over this breach of contract claim under 28 U.S.C. §§ 1332(a) and 1367(a).

ANSWER: Paragraph 84 contains legal conclusions to which no response is required. To the extent a response is required, Counter-Defendants admit that Sarepta's counterclaims purport to assert a breach of contract claim arising under Delaware state law. Counter-Defendants deny

the remaining allegations in paragraph 84.

85. This claim for breach of contract arises out of Nippon Shinyaku's material breach of the MCA with Sarepta.

ANSWER: Counter-Defendants deny the allegations in paragraph 85.

86. Properly interpreted, the MCA is a valid and enforceable contract between Sarepta and Nippon Shinyaku.

ANSWER: Counter-Defendants admit that the MCA is a valid and enforceable contract between Sarepta and Nippon Shinyaku. Counter-Defendants deny any remaining allegations in paragraph 86.

87. Sections 1-3 of the MCA define "Confidential Information" and proscribe improper disclosures or uses of confidential information beyond the permitted purposes. Section 2.2, entitled "Obligations of Confidentiality and Non-Use," states among other relevant provisions that:

> The Parties intend and agree that this Agreement, the Proposed Transaction and all disclosures, including all meetings, discussions, correspondence, communications, documents, or other materials exchanged between the Parties made in connection with this Agreement and the Proposed Transaction shall not be submitted, referenced, admitted or otherwise used by the Recipient, its Affiliates, or their respective Representatives against the other Party in any legal action, except in an action to enforce the terms of this Agreement, and shall be treated as conducted in the aid of negotiation and shall be governed by and entitled to the protections and privileges of Delaware Rule of Evidence 408 and Federal Rule of Evidence 408, as well as any and all analogous or applicable privileges or additional limitations on use and disclosure set forth herein. Furthermore, regardless of whether the Proposed Transaction leads to any arrangement or resolution of issues, the fact that these confidential Proposed Transactions occurred shall not be referenced in any legal action currently pending, including but not limited to the EP Oppositions, the JP Actions, or the Potential Actions. Neither Party nor their Affiliates or Representatives shall in any way attempt to place into evidence any document, fact, statement or opinion in any way relating to the Proposed Transaction for any purpose, regardless of whether such document, fact, statement or opinion would be admissible under FRE 408 or any other analogous or applicable privilege.

D.I. 2-1 at 3.

ANSWER: Counter-Defendants admit that paragraph 87 recites a portion of Section 2.2 of the MCA. Counter-Defendants further admit that the term "Confidential Information" is listed in Section 1 of the MCA along with a definition of the term. Counter-Defendants also admit that the title of Section 2 of the MCA recites "Obligations of Confidentiality and Non-Use." Counter-Defendants deny the remaining allegations of paragraph 87.

88. On July 14, 2021, Nippon Shinyaku filed its original Complaint in this action containing confidential information in violation of its agreement, materially breaching its obligations under the MCA, Sections 1-3.

ANSWER: Counter-Defendants admit that Nippon Shinyaku filed its Original Complaint in this action on July 13, 2021. Counter-Defendants deny the remaining allegations of paragraph 88.

89. Notwithstanding that in its first set of Rule 12 responsive motions Sarepta raised its objection to such confidential information appearing in Nippon Shinyaku's original Complaint contrary to the terms of the MCA, Nippon Shinyaku again included the same confidential material in its First Amended Complaint ("FAC"), filed September 10, 2021 (D.I. 39).

ANSWER: Counter-Defendants admit that Nippon Shinyaku filed its First Amended Complaint on September 10, 2021. Counter-Defendants further admit that on September 3, 2021, Sarepta filed a Motion to Dismiss and Motion to Strike certain paragraphs of the Original Complaint. Counter-Defendants deny the remaining allegations of paragraph 89.

90. Sarepta renewed its objection in subsequent Rule 12 responsive motions (D.I. 53, 54) to such confidential information appearing in Nippon Shinyaku's FAC contrary to the terms of the MCA.

ANSWER: Counter-Defendants admit that Sarepta filed a Motion to Dismiss and Motion to Strike Portions of the First Amended Complaint on September 24, 2021. Counter-Defendants deny the remaining allegations of paragraph 90.

91. On December 20, 2021, the Court found that Nippon Shinyaku had violated the confidentiality and non-use provisions of the MCA and struck from the FAC the second sentence of paragraph 2 as well as paragraphs 11, 78, and 91 of the FAC. (Hearing Tr. at 31-34; D.I. 84.)

ANSWER: Counter-Defendants admit that the Court struck the second sentence of paragraph 2 and paragraphs 11, 78, and 91 of the First Amended Complaint. Counter-Defendants deny the remaining allegations of paragraph 91.

92. As the Court found, Nippon Shinyaku "agreed not to hold the parties' confidential communications against Sarepta in future litigation" because the terms of the valid and enforceable MCA had prohibitions against mentioning confidential communications in legal actions. *Id.* at 32, 34. But Nippon Shinyaku materially breached the terms of the agreement by including confidential information in its original Complaint and again in its FAC, even after being put on notice of its breach, requiring further briefing, motions practice, and a ruling by this Court striking the confidential information from Nippon Shinyaku's pleading.

ANSWER: Counter-Defendants admit that in the Hearing Transcript from the hearing on December 20, 2021, the Court stated that "NS agreed not to hold the parties' confidential communications against Sarepta in future litigation." Counter-Defendants deny the remaining allegations of paragraph 92.

93. Sarepta has suffered prejudice and injury by virtue of Nippon Shinyaku's knowing and repeated bad-faith breaches of the MCA's confidentiality and non-use provisions of Section 2, entitling Sarepta to damages in an amount exceeding $75,000.

ANSWER: Counter-Defendants deny the allegations of paragraph 93.

94. In addition, in view of Nippon Shinyaku's knowing and repeated bad-faith breaches of the MCA, Nippon Shinyaku has unclean hands precluding enforcement of the MCA and depriving it of any entitlement to injunctive or other equitable relief for any alleged breach of the MCA by Sarepta.

ANSWER: Counter-Defendants deny the allegations of paragraph 94.

GENERAL DENIAL

Except as expressly admitted in the preceding paragraphs above, Counter-Defendants deny each and every allegation of Sarepta's Counterclaims including, without limitation, the headings and subheadings contained in the Counterclaims. Pursuant to Rule 8(b) of the Federal Rules of Civil Procedure, allegations contained in the Counterclaims to which no responsive pleading is required and allegations for which Counter-Defendants lack knowledge or information sufficient

to form a belief about the truth of the allegations shall be deemed denied. Counter-Defendants expressly reserve the right to amend and/or supplement their answer.

RESPONSE TO PRAYER FOR RELIEF

Counter-Defendants deny that Counter-Plaintiffs are entitled to the relief it requests or to any other relief.

RESPONSE TO DEMAND FOR A JURY TRIAL

Counter-Defendants admit that Counter-Plaintiffs have demanded a jury trial for all triable issues alleged in its counterclaims but denies that a jury trial is warranted for Counterclaim V.

DEFENSES

Without assuming any burden other than those imposed by operation of law, and without admitting that they bear the burden of proof with respect to any of the following, Counter-Defendants, on information and belief, while reserving the right to add additional defenses based on facts learned in discovery or otherwise assert the following defenses.

First Defense
(Non-Infringement of the UWA Patents)

Counter-Defendants have not infringed and will not infringe, directly or indirectly, any valid and enforceable claim of the UWA Patents, either literally or under the doctrine of equivalents.

Second Defense
(Invalidity of the UWA Patents)

Each asserted claim of the UWA Patents is invalid for failure to comply with one or more requirements of the patent laws of the United States, including without limitation, 35 U.S.C. §§ 101, 102, 103, 112, and/or obviousness-type double patenting, and the rules, regulations, and laws pertaining thereto.

Third Defense
(Prosecution History Estoppel and Disclaimer)

Counter-Plaintiffs' claims that Counter-Defendants infringe the UWA Patents are estopped in whole, or in part, by representations made or actions taken during the prosecution of the applications that lead to the UWA Patents and/or related patents under the doctrine of prosecution history estoppel and/or prosecution history disclaimer.

Fourth Defense
(No Invalidity of the NS Patents)

All claims of the NS Patents are not invalid or unenforceable under 35 U.S.C. § 1 *et seq.*, and Counter-Plaintiffs will not be able to demonstrate otherwise by clear and convincing evidence.

Fifth Defense
(No Breach of Contract)

Counter-Defendants have not breached any contractual obligations under the MCA. To the extent Sarepta asserts a breach of contract claim against Counter-Defendant NS Pharma, NS Pharma was not a party to the MCA.

Sixth Defense
(Failure to State a Claim)

Counter-Plaintiffs' Counterclaims fail to state a claim upon which relief may be granted.

Seventh Defense
(Equitable Defenses and Remedies)

Sarepta's breach of contract claim and/or requested remedies arising from said breach of contract claim are barred in whole or in part under principles of equity, including unclean hands. By way of example only, in light of Sarepta's breach of the MCA by filing its IPR Petitions before the PTAB instead of challenging the validity of the NS Patents in the District of Delaware, Sarepta has unclean hands precluding it from enforcing the MCA and depriving it of any entitlement to injunctive or equitable relief for any alleged breach of the MCA by Counter-Defendants.

Eighth Defense
(No Damages)

Counter-Plaintiffs have not incurred any damages resulting from its allegations that Counter-Defendants have infringed the UWA Patents and/or breached the MCA. Counter-Defendants deny any allegations of infringement of the UWA Patents and breach of the MCA.

Ninth Defense
(Limitation on Damages and Costs)

Counter-Plaintiffs' claims for relief are barred in whole or in part, including without limitation by 35 U.S.C. §§ 286, 287, and/or 288.

Tenth Defense
(No Willful Infringement of the UWA Patents)

Counter-Defendants have not willfully infringed the UWA Patents, and Counter-Plaintiffs are therefore not entitled to enhanced damages pursuant to 35 U.S.C. § 284.

Eleventh Defense
(No Exceptional Case)

Counter-Plaintiffs cannot prove that their case against Counter-Claim Defendants is exceptional and warrants the award of attorney fees under 35 U.S.C. § 285 or pursuant to the Court's inherent power.

Reservation of Additional Defenses

Counter-Defendants reserve the right to add additional defenses based on facts learned in discovery or otherwise.

PRAYER FOR RELIEF

WHEREFORE, Counter-Defendants respectfully request the following relief:

A. A judgment in favor of Counter-Defendants with respect to Counter-Plaintiffs' Counterclaims;

B. A judgment that Counter-Plaintiffs are not entitled to any of the relief requested in their Counterclaims and dismissal of Counter-Plaintiffs' Counterclaims with prejudice;

C. A judgement declaring that, pursuant to 35 U.S.C. § 285, this is an exceptional case and awarding Counter-Defendants their attorneys' fees;

D. A judgement awarding Counter-Defendants their costs under Fed. R. Civ. P. 54(d) and 28 U.S.C. § 1920; and

E. Such other and further relief as the Court may deem just and proper.

Dated: May 5, 2023

Respectfully submitted,

MORGAN, LEWIS & BOCKIUS LLP

/s/Amy M. Dudash
Amy M. Dudash (DE Bar No. 5741)
1201 N. Market Street, Suite 2201
Wilmington, Delaware 19801
Telephone: 302.574.3000
Fax: 302.574.3001
amy.dudash@morganlewis.com

Attorneys for Counter-Defendants Nippon Shinyaku Co., Ltd. and NS Pharma, Inc.

Amanda S. Williamson (admitted *pro hac vice*)
Christopher J. Betti (admitted *pro hac* vice)
Krista V. Venegas (admitted *pro hac* vice)
Wan-Shon Lo (admitted *pro hac vice*)
Maria E. Doukas (admitted *pro hac vice*)
Zachary D. Miller (admitted *pro hac vice*)
Michael T. Sikora (admitted *pro hac vice*)
110 N. Wacker Drive, Suite 2800
Chicago, IL 60601
Telephone: 312.324.1000
Fax: 312.324.1001
amanda.williamson@morganlewis.com
christopher.betti@morganlewis.com
krista.venegas@morganlewis.com
shon.lo@morganlewis.com
maria.doukas@morganlewis.com
zachary.miller@morganlewis.com
michael.sikora@morganlewis.com